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Case report

# Bilaterally impaired hand dexterity with posterior cortical atrophy

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ABSTRACT

A 79-year- old man presented with bilaterally impaired hand movements pertaining to handling of objects although hand movements without the use of objects were preserved, findings consistent with tactile apraxia. His hand and finger movements were slow and clumsy. He had an isolated optic ataxia, a component of Balint's syndrome. The computed tomography scan showed enlargement of the posterior horns of the lateral ventricles. He had recurrent falls probably owing to visual attentional deficits, which may be present in patients with posterior cortical atrophy. The findings can be deemed to fall within the posterior cortical atrophy spectrum. The underlying mechanisms are discussed.
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## 1. Introduction

Klein[1] first used the term “tactile apraxia” to define the loss of purposive movements of the hand. Tactile apraxia is characterized by an isolated disturbance of hand movements for use of and interaction with objects (transitive), whereas intransitive movements are preserved.[2] It could impair hand and arm movements to the same extent as motor impairment resulting from damage to the motor cortex.[3] In patients with tactile apraxia, the lesion involves mainly the posterior parietal cortex, and posterior parietal lesions can give rise to severe disturbances of purposive behavior of the hand during explorative finger movements and manipulation of objects.[3] Posterior cerebral dysfunction may be the result of a variety of causes such as primary degenerative cerebral diseases, vascular causes, tumors, or metabolic factors.

Posterior cortical atrophy (PCA) is a clinicoradiologic syndrome.[4] It is a progressive neurodegenerative disorder characterized by an array of manifestations that include higher visual dysfunction with subsequent development of alexia, agraphia, visual agnosia components of the Balint's and Gerstmann's syndromes, and transcortical sensory aphasia attributed to involvement of the parietal and occipital cortices.[5] Among patients with PCA, several symptom clusters or subsyndromes emerged that have been recognized to have neurological correlates, for instance, Gerstmann's syndrome is attributable to left angular gyrus dysfunction and Balint's syndrome is attributable to bilateral parieto-occipital dysfunction. Memory and cognition are spared until late in the course of the disease.[5] Frontal lobe involvement becomes evident as the disease progresses. We describe a man with bilaterally impaired hand movements, severe ataxia, and recurrent falls associated with PCA.

## 2. Case report

A 79-year-old right-handed man was seen with a history of unsteady gait and recurrent falls, the duration of which was unclear but could be for 1 year or more. When seen he was able to stand on his own and could walk alone but unsteadily. He had mild hypertension and was on enalapril (Renitec; 5 mg daily). Examination of the nervous system revealed no weakness of the limbs. He was seen a few months later when his condition had worsened. His wife volunteered the information that she had to buy him several teacups with different sized handles as he was unable to maneuver his fingers of either hand to hold the cup by the handle. The cranial nerves were intact. Visual fields were normal by direct

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**Table 1**
Neurological findings.

| | |
|---|---|
| Motor | |
| Power | Normal |
| Movements of hands and fingers[a] | |
| 5 finger movements | Reduced, poor sequencing |
| Alternate, repetitive movements | Slow, clumsy |
| Paper crumbling, one finger tapping | Ineffective |
| Explorative pattern | Meaningless, ineffective |
| Reciprocal coordination | Not possible |
| Bimanual tasks—buttoning | Slow |
| Object bound hand movement with visual guidance | Not possible |
| Finger identification | Partially possible |

[a] Bilaterally.

confrontation, pupillary reflexes were normal, and the external ocular movements were omnidirectional. Muscle strength and bulk were equal and normal in all four limbs. The tone was slightly increased in the limbs. The reflexes were normal and the plantars were flexor. Sensations for touch pain, deep pressure pain, and discriminative sensations such as joint sense, topographical localization, and direct simultaneous stimulation were normal. Stereognosis for size and shape was possible for large objects but he had difficulty with small objects because of decreased manipulative hand movements. His memory and cognition were mildly impaired. He has had no formal (school) education. He had never learned to read, write, or spell but could add and subtract single digits. Hence, the following items from the Mini Mental State Examination were not included: to spell, W O R L D and in reverse (5 points), write a sentence (1 point), and read "Close your eyes" (1 point), and his final score was 19 out of 23.

He was unable to direct his hands to targets under visual guidance, for example, when asked to touch a number of targets on a picture he was unable to do so, pointing erratically, nor was he able to trace the outline of the map (optic ataxia). He was able to gaze when desired (no apraxia of gaze) and was able to pick up almost all the objects on a tray (no simultanagnosia). He was unable to perform rapid finger movements with or without visual guidance in both hands. Sequential movement of the fingers—touching the tip of the thumb with the palmar surface of the other four fingers—was not possible in both hands with eyes closed and open. Right–left discrimination was possible, and finger identification was partial. There was no apraxia for intransitive movements such as to wave goodbye, stop, go, and salute, but not to transitive movements such as opening the door with the key, using a scissors, and using a screwdriver. The neurological findings are summarized in Table 1.

The carotid Doppler revealed mildly echogenic plaques in both carotid bulbs at the origin of both internal carotid arteries. The right carotid showed 15–49% stenosis. The left carotid was normal. The computed tomography (CT) scan of the brain showed marked enlargement of the occipital horns of the lateral ventricles and widened sylvian fissures together with prominent sulci over the parietal and occipital regions bilaterally. There were hypodense areas in relation to the anterior horns and in the paraventricular regions. There was no evidence of recent or old cerebral infarction (Fig. 1).

## 3. Discussion

There are several subsets in PCA consisting of variable components of Balint's and Gerstmann's syndromes, but the most

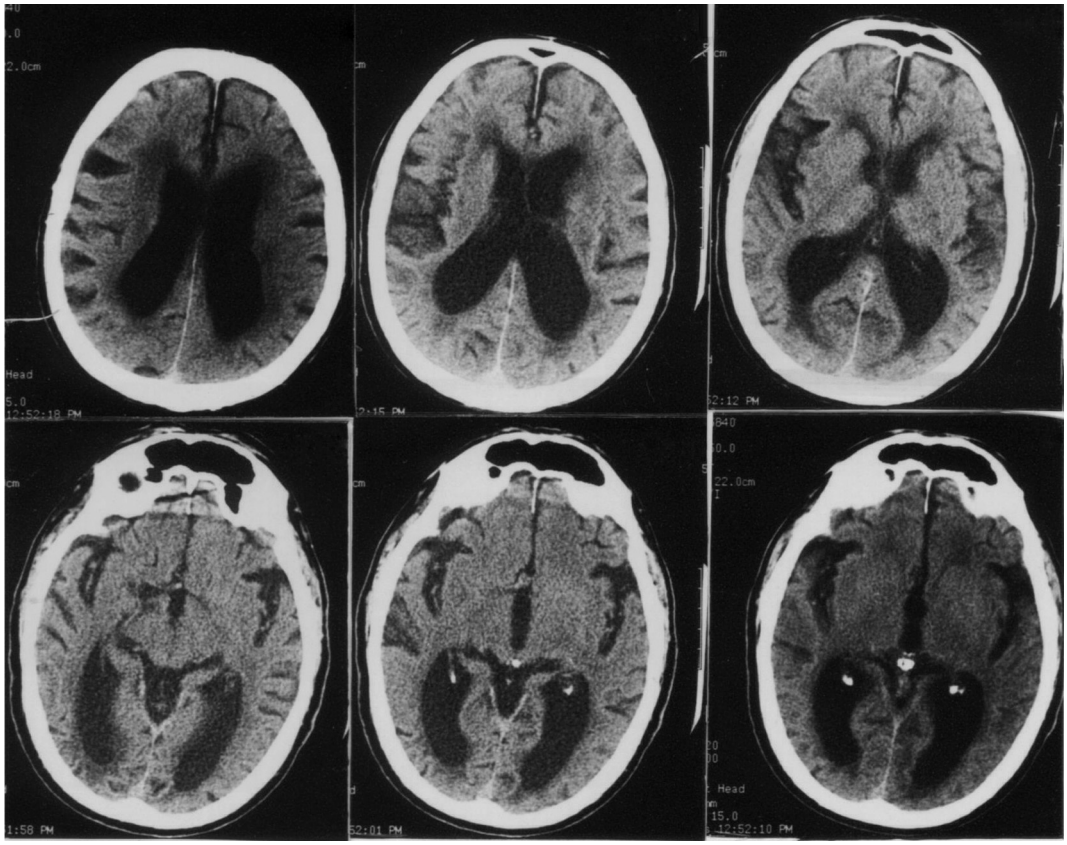

**Fig. 1.** Computed tomography (CT) scan showing bilateral symmetrical striking enlargement of the posterior horns of the lateral ventricles with prominent sulci in the parietal and occipital regions and widening of the sylvian fissures bilaterally.

common neuropsychological symptoms seen are the full Balint's syndrome, which refers to optic ataxia, apraxia of gaze, and simultanagnosia or some component of the syndrome[6] and attributed to visuospatial perception deficits.[7] All of the symptoms are not necessarily present in each case, suggesting that they do not rely on a single brain mechanism.[7] The manifesting component will depend on the structure and function of the area involved. Our patient manifested only one single component (optic ataxia). Careful bedside testing have demonstrated signs of disproportionate parietal/occipital dysfunction.[4] The core features of the syndrome that had been proposed for the diagnosis of PCA are (1) insidious onset and slow progression; (2) visual deficits in the absence of ocular disease; (3) relatively preserved memory, language, and insight; (4) symptoms of Balint's and Gerstmann's syndromes; and (5) absence of tumor or stroke.[4,8,9] The case presented here fits all five criteria.

Optic ataxia may be a rare as an isolated sign or may be a prominent sign or may be missed because of lack of awareness. According to Balint, optic ataxia is a disconnection between visual and motor centers, and gaze apraxia is a consequence of attention impairment.[7] Selective involvement of the dorsal occipitoparietal pathways gives rise to optic ataxia, apraxia of gaze, and asimultanagnosia (Balint's syndrome), and the ventral occipitofugal pathways to agnosia, prosopagnosia, and alexia and involvement of the adjacent inferior parietal lobes to Gerstmann's syndrome. Posterior parietal lesions, in comparison to anterior parietal lesions, exhibit severe astereognosis and apraxia and deficits in dexterity feature a more pronounced decrease in regularity and frequency of manipulative movements.[2]

Both tactile and optic (visuomotor) apraxias are unimodal apraxias where movement disturbance affects only one particular sensory domain, leaving all other aspects of motor behavior intact.[10] Our patient had both tactile apraxia and optic ataxia. Whether both can occur together is a moot point. The motor deficit in patients with optic ataxia (visuomotor apraxia) is similar to that described for patients with tactile apraxia,[3] so much so that Freund[3] regarded tactile apraxia and visuomotor ataxia (optic ataxia) as deficits of sensory-guided motor behavior due to lesions in the posterior cerebral cortex rather than to unimodal differentiation or disconnection syndromes.[3] White matter and cortical changes occur in PCA and, according to Freund,[10] large lesions involving white matter are not easy to elucidate because it is difficult to be certain what functional disturbance comes from white matter or cortical damage.

The syndrome is associated with a variety of pathologies.[4] Alzheimer's disease is the most common underlying pathology, but several workers have associated it to other etiologies such as dementia with Lewy Bodies,[11,12] and corticobasal degeneration.[11,12] Many of these cases displayed neurofibrillary tangles and senile plaques in the parieto-occipital regions, whereas the frontal regions were less affected. Structural neuroimaging demonstrated parieto-occipital atrophy, and functional imaging has revealed bilateral hypometabolism and hypoperfusion in these areas.[13] Diffusion tensor imaging has shown loss of white matter microstructure integrity that increases with age[14] and owing to either white matter lesions or white matter atrophy. Focal cortical atrophy giving rise to a variety of symptoms clusters reflects the distribution of the lesion rather than the pathology. Our patient showed marked dilatation of the posterior horns of the lateral ventricles. Localized ventricular dilatation of the brain can be due to *ex vacuo* dilatation resulting from either cerebral atrophy (gray or white matter or both) or to structural damage to the brain parenchyma as from stroke or trauma.[6] The volume of cerebral ventricles is determined by the nuclei and white matter tracts that adjoin them.[15]

The patient had bilateral incoordination of the hands for simple and complex movements. He had disturbed explorative hand movements (transitive), but expressive movements (intransitive) were preserved satisfying the criteria for tactile apraxia.[1,2] The hands showed clumsiness and slowness. Alternate and repetitive movements and reciprocal coordination were impaired. Limb apraxia had been highlighted as a feature of PCA,[13] and it refers to ideomotor apraxia of the limbs, the hands, and the fingers. A study of 82 patients with “deep” apraxia revealed lesions involving different anatomical sites of importance: the putamen, lenticular nucleus, capsular, periventricular and parastriatal white matter, and the thalamus. And almost in all of them, the lesions encroached into the adjacent lateral white matter to involve the association fibers.[16] A CT scan performed in our patient showed PCA involving the parieto-occipital areas and underlying white matter. There were hypodense areas in relation to the anterior horns and paraventriculuar regions, indicating white matter changes. There was a widening of the sylvian fissures bilaterally. Absence of limb weakness, sensory deficits, and language dysfunction excluded syndromes relating to the precentral and perisylvian regions.

In the PCA syndrome, there may not only be selective involvement of the pathways but beyond what may not be expected from the atrophy. Not only motor deficits can be produced by reduced sensory input to the motor areas but also loss of information from the somatosensory association areas to the areas involved in motor programming.[17] Shibasaki et al[18] has shown that the supplementary motor area (SMA) plays an important role in the planning and execution of simple and complex sequential finger movements. The SMAs are programming areas for motor subroutines. Parietal lesions often cause severe abnormalities of explorative movements and in handling objects during purposive behavior,[3] and the parietal cortex has a prominent role as a sensorimotor interface.[10] Although there was no direct involvement of the SMA, it can be attributed not only to damage to the parietal region but also to the disruption of the occipitoparietofrontal bundle.

An association between handedness, cerebrovascular accident, carotid artery stenosis, and impaired dexterity of the ipsilateral hand has been described.[19] In right-handed patients with left cerebrovascular accident, right hemiplegia occurs but in the presence of a significant right internal carotid stenosis apraxia develops in the left uninvolved hand.[19] Our patient did not have a cerebrovascular accident nor had any carotid artery plaque of any significance.

It has long been known that the ataxia produced by cerebral lesions involving the somatosensory association cortex have a close similarity to that produced by lesions in the lateral cerebellum.[17] According to Montgomery,[17] not only a lesion of the lateral cerebellum would give rise to loss of adequate motor programming and resulting ataxia but also inadequate input from the damaged cerebral part of the cortex (sensory association areas) to the lateral cerebellum. The patient had recurring falls. The ataxic gait could be a risk factor for falls. Visual–spatial attention is one specific aspect of cognition that may be related to falls.[20] Higher-order visual attentional difficulties may be present in patients with PCA,[4] and visual–spatial attention deficit has been related to falls risk.[20]

We have described a 79-year-old man with bilaterally impaired hand movements, unsteadiness of gait, and recurring falls. The CT scan showed PCA with ventricular expansion and atrophy of the adjoining cortices and white matter. The aim was to postulate an association between the presenting symptoms and PCA. The somatosensory association cortices in the parietal lobe involved are the source of information to the SMAs, which play an important role in the planning and execution of simple and complex movements of the hands.[17] Damage to the posterior parietal areas or disruption of the connecting pathways can result in loss of purposive movements of the hands.[3] Reduction of output from the somatosensory areas to

the lateral cerebellum may give rise to ataxia, which may be a risk factor for falls. Alternatively, occipitoparietal regions are associated with visual–spatial integration and attentional mechanisms, and visual–spatial dysfunction has been related to falls risk.[20] With PCA, the symptoms can occur beyond the region of the cerebral atrophy, and a single symptom can arise from many areas that are integrated as in the case just described. Our understanding of the clinical manifestations of PCA may still be incomplete; hence, case reports and case series remain an invaluable source of information in this regard.

## Conflicts of interest

All contributing authors declare no conflicts of interest.

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